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An interesting case of inflammatory myofibroblastic tumor presenting as cholangiocarcinoma

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ABSTRACT

INTRODUCTION: Inflammatory myofibroblastic tumor (IMT) is a reactive or inflammatory state mostly affecting the pulmonary system and commonly occurs in children and young adults. IMT presentation in the hepatic duct bifurcation is very rare and has sporadically been reported before.
PRESENTATION OF CASE: A 12-year-old girl presented with jaundice, pruritus which had begun 5 weeks previously. Ultrasound revealed intrahepatic biliary ductal dilation and an isoechoic 25*30 mm lesion at or near the confluence of the right and left hepatic ducts that were suggestive of a hilar cholangiocarcinoma. Limited resection was decided intraoperatively because the intraoperative frozen section assessment of the CBD, right and left hepatic duct wall samples and porta hepatis lymph nodes was normal. Histologically the tumor proved an inflammatory myofibroblastic tumor (IMT).
DISCUSSION: Almost all patients with resectable IMT should be managed with radical surgical resection or single nonsteroidal anti-inflammatory drugs. In addition, conservative treatments with NSAIDs, corticosteroids or chemotherapeutic agents could not be started in many cases due to the lack of definitive diagnosis of the mass preoperatively. Thus, surgical removal is frequently unavoidable.
CONCLUSION: Biliary IBT is extremely rare and should be considered by all hepatobiliary surgeons dealing with the teens with cholangiocarcinoma, to avoid unnecessary major surgical resections.

1. Introduction

Inflammatory myofibroblastic tumor (IMT) is a reactive or inflammatory state mostly affecting the pulmonary system and commonly occurs in children and young adults [1]. It includes a spectrum of myofibroblastic spindle cells proliferation along with varying amount of inflammatory cell infiltration [2]. IMT was earlier known as inflammatory pseudotumor, which was later coined as IMT(as being a more descriptive name) by Scott et al. in 1988 [3]. IMT also can affects other organs such as the lung, liver, spleen, gastrointestinal tract, genitourinary tract, and heart [4]. However, its presentation in the hepatic duct bifurcation is very rare and has sporadically been reported before. Because of its radiologic and behavior similarities to malignancy, IMT can pose a notable diagnostic effort. A variety of diagnostic terms have been applied to this lesion previously, such as; fibrous xanthoma, plasma cell granuloma, pseudosarcoma, lymphoid hamartoma, myxoid hamartoma, inflammatory myofibrohistiocytic proliferation, benign myofibroblatoma [5]. This work has been reported in line with the SCARE criteria [6].

2. Case report

A 12-year-old girl presented with jaundice and pruritus which had been begun 5 weeks previously. She also had a history of minimal weight loss and anorexia. Her gynaecological history was not significant, and her past medical and surgical history were unremarkable. On physical examination she checked out fine, except for the presence of jaundice and scratch marks. Laboratory analysis showed elevated total bilirubin of 19.3, direct bilirubin of 8.3 mg/dl, Alkaline Phosphatase of 1320 U/L and Amylase of 1794 U/L. Other routine laboratory tests were within normal limits except for a slightly elevated aspartate aminotransferase (150 U/L) and alanine aminotransferase (230 U/L).

Ultrasound revealed intrahepatic biliary ductal dilation and an isoechoic 25*30 mm lesion at or near the confluence of the right and left hepatic ducts. Common bile duct diameter (CBD) was 8 mm. The appearances were suggestive of a hilar cholangiocarcinoma (klatskin tumor). Spleen was 130 mm in maximum bipolar diameter with normal parenchyma echo texture.

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Fig. 1. CHD Mass (blue arrow), Dilated Intrahepatic biliary ducts (green arrow).

Abdominal helical CT confirmed these findings. The mass size was estimated 23*17 mm in CT and there were no additional pathological findings. The para-aortic and paraceliac lymph nodes were normal (Fig. 1).

A CA 19-9 of 493 U/ml was detected in further laboratory analysis.

Subsequently, Endoscopic Retrograde cholangiopancreatography (ERCP) was performed which revealed multiple discontinuous stenoses in both proximal and distal part of CBD that were suggestive for primary sclerosing cholangitis or cholangiocarcinoma. A biliary metallic stent was placed into the common bile duct but bilirubin levels did not drop.

There was no evidence of extrahepatic disease or peritoneal seeding or lymphadenopathy on CT scan. Therefore, the patient underwent exploratory laparotomy with the intention of relieving the biliary obstruction and performing a complete resection.

Surgical exploration, revealed a hard neoplastic mass in the CBD extending proximally towards the porta hepatis. The intraoperative frozen section assessment of the CBD, right and left hepatic duct wall specimens (three separated specimens) and porta hepatis lymph nodes showed no signs of malignancy. Hence, limited hepatic resection was performed. Reconstruction of the biliary tract was performed by the end-to-end anastomosis of a normal-appearing intrahepatic bile duct measured 6 mm in diameter to the remained common bile duct. The T-tube is placed in the CBD, anchored to the abdominal wall, and joined to a closed drainage system.

Histologically, the tumor proved an Inflammatory myofibroblastic tumor (IMT), arising from the bile duct epithelium, composed of monomorphic spindle cells fascicles and chronic inflammatory cells including predominantly lymphocytes and few plasmacells and reactive mesenchymal tissues. IHC study reveal positive staining of spindle cells for SMA, Beta-cathenin and ALK and negative reactivity for caldesmon.

The postoperative course was uneventful (excepting some episodes of low-grade fever). Serum bilirubin and amylase levels were dropped to normal limits within 20 days after the relief of the biliary obstruction. She was discharged 21 days postoperatively.

3. Discussion

Inflammatory myofibroblastic tumor (IMT) has been classified as tumors of intermediate biological potential by World Health Organization (WHO) due to a tendency of local recurrence and a lower risk of distance metastasis [6]. Various theories have been proposed to explain the patogenesis of IMT, such as; reactive etiology, infectious agents or neoplastic processes (cytogeneticaberrations involving the anaplastic lymphoma kinase (ALK) gene on chromosome 2p23, which occur in approximately 50% of IMTs) [7]. The presence of human herpesvirus-8 DNA sequences and the over expression of interleukin 6 and cyclin D1 have been reported in IMTs [8]. IL-6 promotes proliferation of fibroblasts. Major origins of IL-6 are monocytes and macrophages which are constant composition of IMT [9,10]. Due to disease progression, lack of alternative therapeutic regimens, recurrence, almost all patients with resectable IMT have been managed with radical surgical resection or single nonsteroidal anti-inflammatory drugs (NSAIDs) [11]. Non, surgical management of IMT, includes anti-inflammatory agents (like NSAIDs or corticosteroids) and chemotherapeutic agents (like cyclosporine, methotrexate, azathioprine, and cyclophosphamide) [12]. Conservative treatments with NSAIDs, corticosteroids or chemotherapeutic agents could not be started in many cases due to the lack of definitive diagnosis of the mass preoperatively. The outcome of IMT depends on its behavior which ranging from completely benign to malignant with fatal outcome [13].

In our case, IHC study reveal positive staining of SMA, ALK and Beta-cathenin in the spindle-shaped cells.

In conclusion, biliary IMT is extremely rare and frequently difficult to distinguish from hepatic malignant processes due to nonspecific imaging characteristics. IMT should be considered by all hepatobiliary surgeons dealing with the teens with cholangiocarcinoma, to avoid unnecessary major surgical resections. Percutaneous needle biopsy can be performed preoperatively for cases of suspected IMT to obtain a pathologic confirmation and to aid in exclusion of cholangiocarcinoma.

Conflict of interest

None declared.

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Ethical approval

Ethical approval has been exempted from our institution for this case report.

Consent

All authors declare that ‘written informed consent was obtained from the patient’s parents for publication of this report.

Author contribution

This work was carried out in collaboration between all authors. AT designed the study, wrote the protocol, and GS conceived of the study, and participated in its design and coordination and helped to draft the manuscript. MK managed the literature searches, participated in the design of the study and performed the statistical analysis. All authors read and approved the final manuscript.

Guarantor

ADNAN TizMaghz.

Registration of research studies

This case study is a retrospective report without any intervention.

References

[1] M. Bahadori, A.A. Liebow, Plasma cell granuloma of the lung, Cancer 31 (1973) 191–208 (PMID: 4683037).
[2] C.F. Poh, R.W. Priddy, D.M. Dahlman, Intramandibular inflammatory myofibroblastic tumour: a true neoplasm or reactive lesion? Oral Surg. Oral Med. Oral Pathol. Oral Radiol. Endod. 100 (2005) 460–466 (PMID: 16182167).
[3] T. Rahman, J.D. Sharma, M. Krishnatreya, A.C. Kataki, A. Das, Inflammatory myofibroblastic tumor of the upper alveolus: a rare entity presenting as a jaw swelling, Ann. Maxillofac. Surg. 4 (2) (2014) 227–229, http://dx.doi.org/10.4103/2231-0746.147155 (PMCID: PMC4293853).
[4] A. Martín Malagón, E. López-Tomassetti Fernández, I. Arteaga González, A. Carrillo Pallarés, H. Díaz Luis, Inflammatory myofibroblastic tumor of the distal bile duct associated with lymphoplasmacytic sclerosing pancreatitis. Case report and review of the literature, Pancreatology 6 (1–2) (2006) 145–154 (Epub 2005 Dec 13. PMID: 16354963).
[5] S. Palaskar, S. Koshti, M. Maralingannavar, A. Bartake, Inflammatory myofibroblastic tumor, Contemp. Clin. Dent. 2 (4) (2011) 274–277, http://dx.doi.org/10.4103/0976-237X.91787 (PMCID: PMC3276851).
[6] R.A. Agha, A.J. Fowler, A. Saetta, I. Barai, S. Rajmohan, D.P. Orgill, for the SCARE Group, The SCARE Statement: consensus-based surgical case report guidelines, Int. J. Surg. 34 (2016) 180–186.
[7] B.C. Gleason, J.L. Hornick, Inflammatory myofibroblasTIc tumours: where are we now? J. Clin. Pathol. 61 (2008) 428–437.
[8] C.A. Griffin, A.L. Hawkins, C. Dvorak, C. Henkle, T. Ellingham, E.J. Perlman, Recurrent involvement of 2p23 in inflammatory myofibroblastic tumors, Cancer Res. 59 (12) (1999) 2776–2780 (PMID: 10383129).
[9] J.J. Gómez-Román, G. Ocejo-Vinyals, P. Sánchez-Velasco, E.H. Nieto, F. Leyva-Cobián, J.F. Val-Bernal, Presence of human herpesvirus-8 DNA sequences and overexpression of human IL-6 and cyclin D1 in inflammatory myofibroblastic tumor (inflammatory pseudotumor), Lab. Invest. 80 (2000) 1121–1126 (PMID: 10908158).
[10] C.M. Coffin, J. Watterson, J.R. Priest, L.P. Dehener, Extrapulmonary inflammatory myofibroblastic tumour (inflammatory pseudotumour): a clinicopathologic and immunohistochemical study of 84 cases, Am. J. Surg. Pathol. 19 (1995) 859–872 (PMID: 7611533).
[11] S. Palaskar, S. Koshti, M. Maralingannavar, A. Bartake, Inflammatory myofibroblastic tumor, Contemp. Clin. Dent. 2 (4) (2011) 274–277, http://dx.doi.org/10.4103/0976-237X.91787 (PMCID: PMC3276851).
[12] Y.-L. Tao, Z.-J. Wang, J.-G. Han, P. Wei, Inflammatory myofibroblastic tumor successfully treated with chemotherapy and nonsteroidals: a case report, World J. Gastroenterol.: WJG 18 (47) (2012) 7100–7103, http://dx.doi.org/10.3748/wjg.v18.i47.7100 (PMCID: PMC3531700).
[13] A.W. Shek, P.C. Wu, N. Samman, Inflammatory pseudotumour of the mouth and maxilla, J. Clin. Pathol. 49 (1996) 164–167 (PMID: 8655685 PMCID: PMC500352).